PedsCases Podcast Scripts

This is a text version of a podcast from Pedscases.com on "**Approach to Purpura.**" These podcasts are designed to give medical students an overview of key topics in pediatrics. The audio versions are accessible on iTunes or at www.pedcases.com/podcasts.

Approach to Purpura

Developed by Samantha Lam and Dr. Melanie Lewis for PedsCases.com.
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Introduction

Samantha: Welcome to PedsCases and our series on "Approach to Rashes" My name is Samantha Lam, a medical student from the University of Alberta, and today my guest is Dr. Melanie Lewis, a general pediatrician and professor at the University of Alberta. We will be discussing an "Approach to Purpura" in children. Let's start with a clinical case.

Dr. Lewis: You are a clinical clerk working in the emergency room when a father brings his daughter in because she became increasingly lethargic over the past 24 hours. She had been sick with nausea and vomiting, and developed a rash.

Samantha: Given this story, as a student, what do you expect to find on exam?

Dr. Lewis: When you see her in the ER, she looks quite sick. She appears to be quite sleepy and does not respond to your questions and examination. You immediately notice a non-blanchable violaceous spot on her torso, and her limbs were mottled. You observe that her vitals show a heart rate of 150, a blood pressure of 90/50, a respiratory rate of 35 and a temperature of 40C.

Samantha: Students, how worried are you and what is your management plan? Let's explore this question.

It's easy to brush off a rash since they are common and often benign. But with purpura, you must always rule out potentially dangerous life threatening clinical conditions. The objectives of this PedsCases podcast are to: 1) learn how to identify and describe a purpuric rash; 2) develop an approach to a making a differential diagnosis to a purpuric rash; and 3) learn to identify common, benign, and dangerous presentations of a purpuric rash and 4) describe the corresponding basic management.

Definition of Purpura

Samantha: Let's start with the basics; Dr. Lewis, what is purpura?

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Dr. Lewis: Purpura refers to a non-blanchable purple rash caused by a hemorrhage into the skin and mucosal membranes, which is the reason why this rash is non-blanchable. Since the problem is with bleeding, think of the problem as either a disruption to hemostasis or the vasculature. The lesion is described differently depending on size: petechiae if it is less than 4 mm, purpura if it is between 5 mm to 9 mm, and ecchymoses it is more than 10mm[1].

Samantha: How would a student approach the differential diagnosis in a child with purpura?

Dr. Lewis: One way to form a differential diagnosis is to consider the clinical presentation of the child and appearance of the rash. A sick or febrile child hints at something serious that needs immediate management, such as infection, disseminated intravascular coagulation, and hemorrhage, while a well appearing child with purpura may indicate vasculitis, thrombocytopenia, or mechanical causes of bleeding. Next, determine if the purpura is palpable.

History and Physical Exam

Samantha: What are key questions to figuring out what is causing purpura?

Dr. Lewis: Go back to the basics: history and physical exam. Immediately look to see if the child is sick and requires immediate care. A child is sick if they are unstable and have abnormal vital signs, constitutional symptoms, altered level of consciousness, increased work of breathing or poor perfusion. Get help if you see these signs. If not, take a history and figure out the onset of the rash, course of illness, and baseline health of the patient. Questions about travel, environmental exposures, vaccination status and sick contacts can point to infection. Ingestion of certain medications or drugs can suggest a potential reaction or bleeding. A family history of a bleeding disorder can indicate an inherited bleeding disorder. It is critical to know if there are constitutional symptoms. Fever and lethargy point to an infection, while weight loss, bone pain, joint pain, pallor, lymphadenopathy and splenomegaly suggest a malignancy.

Samantha: Are there any specific physical exam findings that students' should look for that can help form their differential?

Dr. Lewis: Start with vitals, vitals, vitals! And assess their circulation, airway and breathing. On the physical exam, expose the child to inspect the entire skin surface. Describe any rash you find using the SCALDA acronym: size, color, arrangement, lesion morphology, distribution, and adjunct structures. Do not forget to look behind the ear, in the pelvic region and at the mucous membranes. Specifically, look for lymphadenopathy, splenomegaly and pain to the joints and bones.

Samantha: Especially with a history with vague unspecific symptoms, a thorough physical exam is important to forming a differential.

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[1]Piette, W. 2012. Pupura. Bolognia. J, Jorizzo. J, and Schaffer. J. Dermatology. 357-368. Elsevier.

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Differential Diagnosis

Samantha: Now, let's start with a presentation that's common in children but non-life threatening.

Dr. Lewis: Henoch-Schonlein Purpura or HSP is a common childhood vasculitis, most commonly occurring in children ages 3 to 15 years of age. The key feature is palpable purpura. The rash is usually below the waist in the legs and buttocks. The child may also have systemic symptoms including abdominal pain, polyarthralgia and signs of renal disease such as hematuria or hypertension. Some children may have a low grade fever[2]. The rash is often the first sign of HSP and will last around 1 week. Abdominal pain may occur 1 week before or after the onset of the rash. Platelet counts will be normal. But half of the cases will have renal complications ranging from asymptomatic microscopic hematuria to acute or chronic renal failure[3]. HSP is usually self-limiting in 3-4 weeks and management consists usually of symptomatic relief[4]. NSAIDs and prednisone may be considered if there is significant joint involvement and abdominal pain[5].

Samantha: With HSP, this self-resolving vasculitis is usually not concerning and management is conservative with blood pressure and urinalysis surveillance for renal complications. What other diseases are also less concerning?

Dr. Lewis: Immune thrombocytopenia or ITP is one of the most common causes of thrombocytopenia in kids The most common age groups affected are between the ages of 1-5 years and are otherwise healthy children present who then have sudden onset of petechiae, purpura, or ecchymoses[6]. ITP commonly present following a recent history of a viral infection Children can also have nosebleeds, bloody stools or heavier menses. ITP may be primary or secondary, but all have low platelet counts. Most children will recover within 3 months of presentation, even without treatment. In the few with severe bleeding such as heavy epistaxis, GI bleeding, or intracranial hemorrhage, they should be admitted into hospital for stabilization and treatment with IVIG or anti-D immunoglobulin, or glucocorticoids. Overall, these children should avoid vigorous physical activity[7].

Samantha: Why is it important to determine if a child with purpura is febrile?

Dr. Lewis: As a general consideration for a child with fever and a purpuric rash, sepsis must be considered in your differential. A septic child looks sick and has abnormal vital signs: fever, tachycardia, tachypnea and in the late stages, hypotension. Your priority is to

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[2] Foreman J, Jolanda van Zuuren. E, and Ehrlich A. Henoch-Schonlein Purpura. Dynamed. 2015.

[3] *Ardoin. S and Fels. E.* 2016.Henoch-Schonlein Purpura. Kliegman.R, Stanton. B, *et al.* Nelson Textbook of Pediatrics. 1216. Elsevier.

[4] Dedeoglu.F, and Kim.S. Henoch-Schönlein purpura (immunoglobulin A vasculitis): Clinical manifestations and diagnosis. Sundel.R, TePas.E. UpToDate. 2015. Wolters Kluwer.

[5] Dedeoglu.F, and Kim.S. Henoch-Schönlein purpura (immunoglobulin A vasculitis): Management. Sundel.R, TePas.E. UpToDate. 2015. Wolters Kluwer.

[6] *DynaMed* [Internet]. Ipswich (MA): EBSCO Information Services. 1995 – 2016. Immune Thrombocytopenia; [updated 2015 Aug 25; cited 2016 Mar 23];. Available from http://search.ebscohost.com.login.ezproxy.library.ualberta.ca/login.aspx?direct=true&db=dme&AN=114263&site=dynamed-live&scope=site. Registration and login required.

[7] Bussel, J. Immune Thrombocytopenia in Children: Initial Management. Mahoney.D, Armsby.C. UpToDate. 2015.

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stabilize the child's ABC- airway, breathing, and circulation, and if possible, do a full septic work up. In children, the infection you must rule out is meningococcemia, which is caused by *Neisseria meningitides.* Affected children are usually younger than 5 years, late adolescent or young adults, but any age group can be affected. Meningococcemia is rapidly fatal if not treated with empiric antibiotics and supportive therapy. Children initially present with non-specific flu-like symptoms and fever before the signs and symptoms of sepsis appear[8]. The petechiae often present on the trunk and extremities, but can be found anywhere on the body, like the mucosal membranes, head, palms, and soles. The centre of the petechiae may transform into a grey color and then become necrotic. In the severest form, the petechiae become palpable and purulent[9]. Children with meningococcemia are often admitted to the PICU because of the severity of their clinical status. They often need significant fluid resuscitation, inotropes and antibiotics. A definitive diagnosis is made via positive blood and/or CSF cultures; but again, do not delay antibiotics if the patient is unwell and unstable.

Samantha: With meningococcemia being so dangerous, what other infections are a cause for concern for a patient presenting with purpura and fever?

Dr. Lewis: Hemolytic uremic syndrome or HUS which may also present with a fever and purpuric rash. HUS classically presents with the triad of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury, and is commonly caused by the ingestion of *E. coli* 0157H7. Children often present with a one week history of fever, and GI symptoms: often bloody diarrhea accompanied by nausea and vomiting. They may look lethargic, confused, and complain of abdominal pain. Purpura and/or petechiae may be present. These children usually require supportive therapy and antibiotics should be avoided. Meticulous fluid management is required as HUS can lead to acute kidney injury and renal failure, and even some may require dialysis. Blood transfusions may be necessary for children with severe anemia[10,11].

Samantha: We have learned that infections causing meningococcemia, HUS, and septic shock are acute life threatening presentations of fever and purpura. Are there any other presentations that may be related to these causes?

Dr. Lewis: Disseminated intravascular coagulopathy or DIC. It is a syndrome that presents acutely in sick patients who have fever, hypoxia, hypotension and signs of shock. Hemorrhage is the most common form of bleeding, followed by purpura and purpura fulminans. DIC is due to hemorrhage and microvascular thrombosis, and is secondary to an underlying cause, like infection, cancer, trauma, and toxic exposures, hence the underlying cause must be considered and treated. Bleeding commonly occurs at venipuncture sites,

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[8] Apicella. M. Clinical manifestations of meningococcal Infection. Calderwood. S, Edwards. M, and Baron. E. UpToDate. 2015. Wolters Kluwer.

[9] *DynaMed* [Internet]. Ipswich (MA): EBSCO Information Services. 1995 – 2016. Meningococcal Disease; [updated 2016 Mar 16; cited 2016 Mar 23]; Available from http://search.ebscohost.com.login.ezproxy.library.ualberta.ca/login.aspx?direct=true&db=dme&AN=116912&site=dynamed-live&scope=site. Registration and login required.

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[11] Niaudet.P. Overview of Hemolytic Uremic Syndrome in Children. Mattoo.T, and Kim. M. *UpToDate.* 2015. Wolters Kluwer.

but may manifest anywhere on the body. Patients' require hospital admission to address the underlying etiology of the DIC and the provision of replacement therapy for the consumption of platelets and clotting factors[12, 13].

Samantha: So far we have discussed acute onset of common, benign, and dangerous causes to a purpura. Are there any causes that are more insidious in terms of their onset?

Dr. Lewis: Childhood cancers are rare, but when they occur, the cancer usually involves the lymphohematopoietic system. Leukemia and lymphoma can present with fever and a purpuric rash. Cancers are hard to detect because the symptoms are often nonspecific and vague. Children often present with constitutional symptoms such as recurrent fevers, night sweats, and weight loss.. As the cancer progresses, coagulopathies and thrombocytopenia occur leading to easy bruising, bleeding, petechiae and purpura. The CBC and differential and peripheral smear will show abnormalities and imaging may show masses. Once you suspect malignancy, a pediatric oncologist should be consulted[14, 15, 16].

Samantha: A large part of our discussion has been focused on fever and purpura with acute and insidious onset that can be life-threatening. Are there any other causes of purpura that require a high degree of suspicion, but may be overlooked?

Dr. Lewis: Whenever you note bruising, you should always consider inflicted trauma or child abuse in your differential, you should suspect abuse from inflicted trauma if the purpuric rash is found in unusual spots or is incongruent to the patient's story, development, and absence of medical issues. Pattern bruises such as in the shape of a hand print, fingers, or objects are obviously concerning for inflicted trauma and require an explanation. Other bruising that is concerning inflicted trauma include bruising on posterior surface, bilateral or symmetric bruising, bruising on ears, and any bruising on a young infant; there is an excellent saying, "if you aren't cruising, you aren't bruising" are suspicious of being inflicted. Any bruising on a young infant requires an explanation. If you suspect child abuse from the history and physical exam, you need to notify child protective services[17].

Conclusion

Samantha: Today, we have reviewed an approach to purpura in children. The key point to this podcast is differentiating a child who is sick with purpura and one who is relatively well.

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[12] *DynaMed* [Internet]. Ipswich (MA): EBSCO Information Services. 1995 – 2016. Disseminated Intravascular Coagulopathy; [updated 2016 Mar 16; cited 2016 Mar 23]; Available from http://search.ebscohost.com.login.ezproxy.library.ualberta.ca/login.aspx?direct=true&db=dme&AN=116912&site=dynamed-live&scope=site. Registration and login required.

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[15] Neville.K, and Steuber. C. Clinical assessment of the child with suspected cancer. Pappo.A, and Armsby.C. UpToDate. 2016. Wolters Kluwer.

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Dr. Lewis: Remember, if the child has fever and purpura, a bacterial infection such as meningococcemia or strep pneumoniae must be considered in your differential diagnosis. Another acute presentation is DIC, where the sick child will have purpura and hemorrhage, and will need to be treated for the underlying cause, thrombocytopenia, and depleting clotting factors. HUS may present with purpura, but patients will have the classic triad of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury along with a common history of bloody diarrhea. These children require supportive therapy and vigilant fluid management.

Dr. Lewis: In the realm of dangerous presentations of purpura, do not forget about the potential for inflicted trauma, especially if the story and bruising pattern are suspicious for abuse. The child's safety is priority and child protective services need to be notified.

Samantha: A more insidious presentation of dangerous purpura is childhood cancer. The presentation may include fever of unknown origin, constitutional symptoms, and easy bruising. These children may have thrombocytopenia along with other abnormalities in the complete blood count, including low or high WBC and anemia. Urgent consultation with a pediatric oncologist is warranted.

Dr. Lewis: Common things being common, Henoch-Schonlein Purpura is the most common vasculitis in children that presents with palpable purpura over mainly the lower extremities and usually has a self-limiting course. Don't forget to check blood pressure and complete a urinalysis to assess for renal involvement. Immune thrombocytopenia or ITP presents in a healthy looking child with unexplained isolated petechiae, purpura, or ecchymoses which may require treatment depending on the degree of thrombocytopenia and bleeding.

Samantha: Thank you for joining this edition of PedsCases on an approach to purpura. Please visit our other podcast series on rashes to explore other rash presentations.

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